BRIEF COMMUNICATIONS

DOUBLE-OUTLET RIGHT ATRIUM WITH RESTRICTIVE OSTIUM PRIMUM AND INCOMPLETE SUPRAVALVULAR RING PRESENTING AS CONGENITAL MITRAL VALVE STENOSIS

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We have recently encountered a rare case of partial atrioventricular (AV) canal. The malformation included a persistent left superior vena cava draining into a dilated coronary sinus. The left and right auriculoventricular valves were abnormal, as usually seen in partial AV canal. The restrictive ostium primum and surrounding atrial septum were malaligned and displaced in a left posterior position. An incomplete supravalvular ring was found above the lateral component of the left AV valve. This anomaly, which is identified as double-outlet right atrium, was misdiagnosed before the operation and was responsible for a clinical presentation similar to congenital mitral valve stenosis resulting from supravalvular ring.

We present a case to draw attention to this rare diagnosis (double-outlet right atrium) as an unusual cause of impediment to left AV valve inflow and to include it in the differential diagnosis of mitral valve stenosis with basically normal subvalvular apparatus.

The patient was a 14-year-old girl, known since childhood to have a cardiac murmur. Although she had reduced physical activity, she developed satisfactorily, without cyanosis. When she was 8 years old, a diagnosis of congenital mitral stenosis resulting from a supravalvular ring was made, and it was confirmed 2 years later by a different pediatric cardiologist. In June 1993, surgical repair was advised owing to cardiac enlargement. The New York Heart Association functional class was II. On physical examination, the patient was a thin prepubertal noncyanotic girl with mild psychomotor impairment. The sternum was of pectus excavatum type. The jugular venous pressure was within normal limits. No peripheral edema was apparent. Cardiac auscultation revealed a holosystolic grade 3/6 murmur, best heard at the apex and the axillar area. There was also a grade 2/6 diastolic murmur. The second heart sound was normal. The lungs were clear to auscultation, and the liver and spleen were not enlarged.

The electrocardiogram showed normal sinus rhythm with electrical signs of right ventricular hypertrophy. The cardiothoracic ratio was 0.55, and the aspect of the left ventricle was suggestive of mitral stenosis. The preoperative echocardiogram showed a partial AV canal type mitral valve with a supravalvular membrane. The left atrium was distended, and moderate mitral valve insufficiency was evident.

At operation, inspection of the heart and great vessels showed a persistent left superior vena cava, which was controlled. Cardiopulmonary bypass was established between the right superior and inferior venae cavae and the ascending aorta. The left atrium was opened parallel to the interatrial groove and revealed frank deviation of the interatrial septum resulting in poor visualization of the mitral valve apparatus. The right atrium was opened and clearly revealed the malalignment of the atrial septum with an ostium primum that bridged the left auriculoventricular orifice. A supravalvular membrane was situated above the anterolateral commissure, the lateral leaflet, and the posterolateral commissure (Fig. 1). The mitral valve was a trileaflet valve, as usually found in partial AV canal. The ostium primum was restrictive and its diameter was approximately 1 cm. The mitral valve was tested and found to be incompetent in its septal commissure (so-called "cleft"). This area was closed with interrupted sutures, leaving a residual orifice of 2.5 cm^2. The malaligned interatrial septum was largely excised and a septal pericardial patch (glutaraldehyde treated) was placed per the usual technique for ostium primum defects. Because of the drainage of the left superior vena cava into a dilated coronary sinus, the coronary sinus was left in its original (right-sided) position to avoid significant right-to-left shunt. The patient was easily weaned from bypass in sinus rhythm, and pressures measured in the left atrium, left ventricle, and ascending aorta were within normal limits. The differentials between the left atrial and diastolic left ventricular pressures and between the aortic and systolic ventricular pressures were also within normal limits.

The postoperative course was uneventful. The postoperative echocardiogram showed satisfactory mitral valve function, without stenosis and with trivial regurgitation.

Malalignment of the atrial septum allowing the right atrium to be connected to both AV valves was first described by Van Mierop[1] in a case of AV septal defect. Few additional cases have been reported since then, almost always associated with AV canal type defects.[2-4] These authors have essentially emphasized the repair technique, which proves to be satisfactory in most instances. This case report shows that this malformation, which is anatomically different from cor triatriatum, should be included in the differential diagnosis of congenital mitral valve stenosis whenever the ostium primum is

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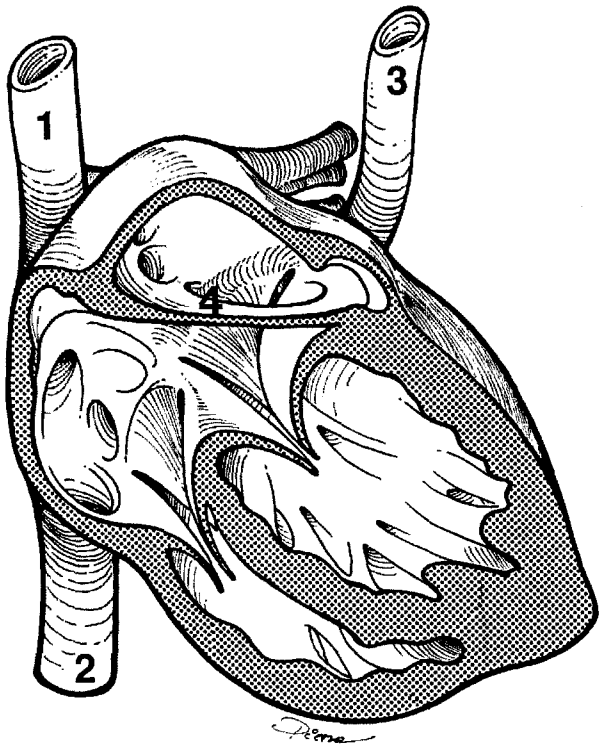

Fig. 1. General appearance of the malformation in the frontal view. *1*, Superior vena cava; *2*, inferior vena cava; *3*, left superior vena cava; *4*, malaligned atrial septum and ostium primum defect.

restrictive and accounts for elevation of left atrial pressure. In accordance with Carpentier and colleagues,[5] when the papillary muscles are basically normal, congenital mitral valve stenosis may be due to papillary muscle commissure fusion, excess valvular tissue, hypoplasia of the anulus, or supravalvular ring. In this case, the supravalvular ring was incomplete, and most of the stenosis was due to the overriding restrictive ostium primum.

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CONGENITAL TOTAL ABSENCE OF THE PERICARDIUM: CASE REPORT OF A 72-YEAR-OLD MAN AND REVIEW OF THE LITERATURE

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Congenital absence of the pericardium (CAP) is rarely encountered during clinical practice. Most patients remain free of symptoms, and the diagnosis is made intraoperatively or at postmortem examination.[1] Some cases have been diagnosed by various imaging techniques, ranging from plain x-ray films to nuclear magnetic resonance. We report the case of a man with coronary artery disease (CAD) who underwent stress thallium scan. The scintigraphic images showed abnormal left ventricular (LV) orientation and levorotation. Intraoperatively, CAP was diagnosed.

Case report. This 72-year-old man with hypertension reported episodic chest pain, dyspnea, and palpitations for

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